

# Pulmonary Artery Stenosis

## A Frequent Part of the Congenital Rubella Syndrome

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■ *The case records, cardiac catheterization and angiographic findings in 32 patients were reviewed to illustrate the high incidence of pulmonary artery stenosis in congenital rubella syndrome. These interesting lesions often are of equivocal or unknown clinical significance compared with the other malformations that beset these patients, but to know of them explains certain physical findings and to study their incidence is an unusual opportunity to pinpoint a definite cause for a congenital cardiac lesion.*

CAMPBELL<sup>2</sup> HAS SAID that patent ductus arteriosus is the most common cardiac malformation met in congenital rubella syndrome. Recently Rowe<sup>8</sup> and Emmanouilides<sup>5</sup> have reported an equally frequent finding, stenosis of the main pulmonary artery branches. Either one or both pulmonary arteries may be discretely narrowed, usually at their origin from the main trunk, and the lesions may occur by themselves or accompanying other cardiac defects. These are uncommon lesions within the population of patients with congenital heart disease,<sup>4,7</sup> but within the population of patients with congenital heart disease due to maternal rubella infection, pulmonary artery stenosis seems to be common. This report describes this coincidence from the experience of the cardiac catheterization laboratory of a general hospital gained through studies on such patients with congenital rubella and pulmonary artery branch stenosis.

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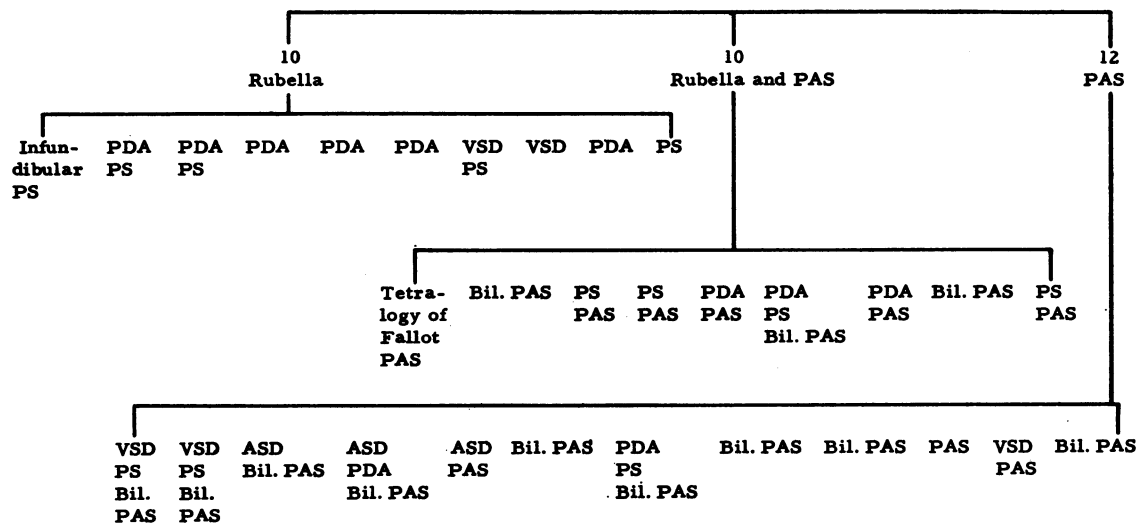
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### Materials and Methods

During the eight-year period 1958-1966, 821 infants, children, or young adults were referred to this center because of a suspicion of congenital heart disease. Twenty of them were regarded as having congenital rubella syndrome because there was a history of maternal rubella infection during pregnancy or there were grounds for suspecting such infection may have occurred and the patient had other congenital anomalies such as cataracts, deafness, mental retardation, microcephaly or microphthalmia.

Also 22 of the 821 patients had pulmonary artery stenosis either as an isolated lesion or combined with other cardiac defects. The diagnosis of pulmonary artery stenosis was based on hearing a systolic murmur originating in the pulmonic area and radiating to the axillae and both sides of the back, recording a sharply localized pressure gradient on pull-through tracings from within the main pulmonary artery branches, and seeing a narrowing on the pulmonary angiogram. In most instances all these conditions were met, but in some cases where cardiac catheterization seemed unnecessary or inadvisable, the diagnosis was based



PS - Pulmonary Valvular Stenosis  
 PAS - Pulmonary Artery Stenosis  
 Bil. PAS - Bilateral Pulmonary Artery Stenosis

PDA - Patent Ductus Arteriosus  
 ASD - Atrial Septal Defect  
 VSD - Ventricular Septal Defect

Chart 1.—Cardiovascular lesions in 32 patients with congenital heart disease.

### 32 PATIENTS

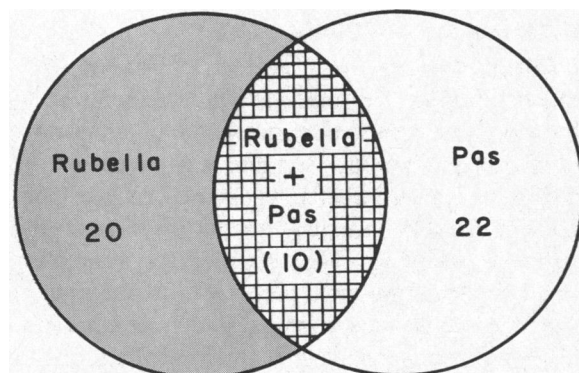


Chart 2.—Overlap between patients having congenital rubella syndrome and pulmonary artery stenosis.

solely on physical findings. Ten of these 22 patients were also in the group having congenital rubella syndrome (Charts 1 and 2).

### Results

Twenty patients had congenital rubella syndrome (Tables 1 and 2). Ten of these had stenosis at one or more sites in the main pulmonary artery branches. There were 12 other patients with pulmonary artery stenosis who had no history or evidence of congenital rubella (Table 3). Accom-

panying cardiac lesions in the patients with rubella syndrome included patent ductus arteriosus (nine cases), pulmonary valvular stenosis (two cases), pulmonary infundibular stenosis (two cases), ventricular septal defect (two cases), and tetralogy of Fallot (one case). Accompanying cardiac lesions in the patients without rubella syndrome included atrial septal defect in three, patent ductus arteriosus in two, pulmonary valvular stenosis in two, ventricular septal defect in one, and acyanotic tetralogy of Fallot in two.

Seven of the patients with bilateral pulmonary artery stenosis showed a characteristic pulmonary trunk pressure curve described by Agustsson and coworkers<sup>1,3</sup> (Figure 1).

In general the stenotic lesions of the pulmonary arteries presented no urgent clinical problems. Surgical repair was attempted in two cases, successfully in a four-year-old girl (Case 13) who benefited from patch grafting of the right pulmonary artery and pulmonary valvotomy, and unsuccessfully in a two-year-old boy (Case 30) with hypoplastic pulmonary arteries and a pressure gradient of 85 mm of mercury between the branches and the main pulmonary artery trunk.

On physical examination the usual finding was a grade II to IV ejection systolic murmur in the

second to third intercostal space to the left of the sternum with wide radiation to both axillae and over the posterior chest to both the right and left sides. In patients with isolated stenotic lesions the second sound in the pulmonic area was either nor-

mal or moderately split—0.04 to 0.06 seconds—and varied with respiration (Figure 2). The electrocardiograms were consistent with incomplete and complete right bundle branch block and right ventricular hypertrophy. Often an associated le-






TABLE 1.—*Congenital Rubella Syndrome without Pulmonary Artery Stenosis*

Case No.	Age at First Visit	Sex	Cardiac Defects	Other Defects	Remarks
1 .....	13 years	F	Infundibular Pulmonary stenosis	Deafness Mental Retardation	.....
2 .....	3 years	F	PDA PS	Bilateral Cataracts	PDA ligated at age 4
3 .....	3 years	M	PDA PS	Bilateral nerve deafness	PDA ligated at age 2 months
4 .....	1 year	M	PDA	Bilateral cataracts & deafness	PDA ligated at age 1 year
5 .....	10 days	M	PDA	Convulsive disorder	PDA ligated at age 10 days
6 .....	4 months	F	PDA	Convulsive disorder, bilateral cataracts, microcephaly	.....
7 .....	2 months	M	VSD PS	Cataract O.D.	.....
8 .....	3 years	F	VSD	Microphthalmia O.S.	VSD repaired at age 3 years
9 .....	10 months	M	PDA	Mental retardation	.....
10 .....	5 months	F	Probable infundibular Pulmonary stenosis	.....	.....

PAS=Pulmonary artery stenosis.  
PS=Pulmonary valvular stenosis.  
PDA=Patent ductus arteriosus.

ASD=Atrial septal defect.  
VSD=Ventricular septal defect.

TABLE 2.—*Congenital Rubella Syndrome with Pulmonary Artery Stenosis*

Case No.	Age at First Visit	Sex	Cardiac Defects	Other Defects	Pressures mm Hg			Angiogram
					MPA	RPA	LPA	
11 .....	6 years	F	Bilateral PAS	Deafness	43/7	16/8	15/7	
12 .....	2 years	F	PS PAS	Deafness Clubbed feet	.....	.....	.....	.....
13 .....	4 years	F	PS PAS	Bilateral nerve deafness	70/14	27/14	.....	
14 .....	2 months	F	PDA PS	Cataract O.S.	.....	.....	.....	.....
15 .....	3 years	F	PDA PAS Aberrant right subclavian	.....	33/10	15/10	.....	.....
16 .....	2 years	F	PDA PS Bilateral PAS	Cataracts & deafness	37/9	19/13	18/11	
17 .....	9 days	F	.....	.....	.....	.....	.....	
18 .....	11 years	F	Bilateral PAS	.....	53/6	20/9	20/10	
19 .....	3 months	F	PAS	.....	.....	.....	.....	.....
20 .....	10 years	M	Tetralogy of Fallot PAS	Deafness	.....	.....	.....	.....

REMARKS—TABLE 2

12. Physical findings characteristic with Gr. II ejection systolic murmur radiating to both axillae and right back.
13. Repair coarctation right pulmonary artery and pulmonary valvotomy at age 4.
14. PDA ligated at age 3 months, systolic murmur radiating over the back persisted.
15. PDA and aberrant right subclavian artery ligation.
17. Ligation PDA at 2 weeks of age.
19. Physical findings characteristic with Gr. II ejection systolic murmur radiating to both axillae and back.
20. Open repair of tetralogy of Fallot at age 4. Persisting systolic murmur radiating to both axillae and back.

sion produced the pattern of biventricular enlargement. X-ray films of the chest were also nonspecific, showing either cardiac enlargement or cardiac

enlargement with prominence of the main pulmonary artery.

## Discussion

After reviewing his own experience and that of others, Campbell<sup>2</sup> decided that between 1 and 2 per cent of congenital heart disease was caused by congenital rubella syndrome. The incidence in the present series (20 of 821 patients) was 2.4 per cent, which is in fairly close accord. D'Cruz and coworkers<sup>3</sup> reported 84 of 2,000 patients (4.2 per cent) were found to have stenosis of one or several areas in the main pulmonary artery or its

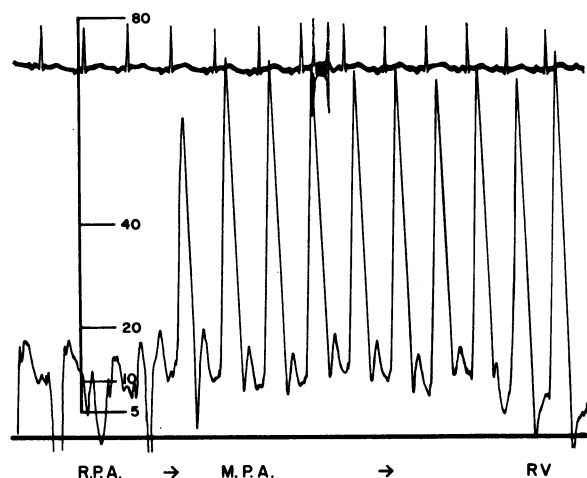


Figure 1.—(Case 18 in Table 2). Pressure curves recorded as the catheter traversed the left pulmonary artery and main pulmonary trunk to the right ventricle. The patient was an 11-year-old girl with bilateral pulmonary artery stenosis.

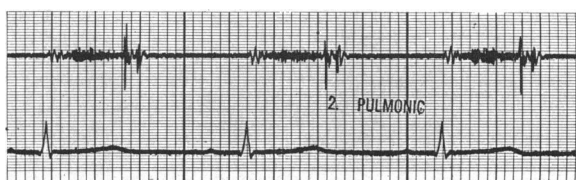


Figure 2.—Phonocardiogram recorded from the pulmonic area in patient with bilateral pulmonary artery stenosis (Case 18, Table 2). There is a systolic ejection murmur and the second sound is split 0.06 second.

TABLE 3.—Pulmonary Artery Stenosis without Congenital Rubella Syndrome

Case No.	Age at First Visit	Sex	Cardiac Defects	Pressures mm Hg			Angiogram	Remarks
				M.P.A.	R.P.A.	L.P.A.		
21	7 years	M	ASD, partial anomalous pulmonary venous drainage to innominate vein, bilateral PAS	43/9	27/10	.....		ASD repaired at age 8
22	7 years	F	ASD, PDA, bilateral PAS	45/17	28/11	.....		PDA ligated at age 2
23	11 years	F	ASD	32/10	18/10	.....	.....	ASD repaired at age 11
24	4 years	F	Bilateral PAS	30/13	17/12	16/12	.....	.....
25	5 years	F	PDA, PS, bilateral	80/15	22/14	.....		Ligation PDA, pulmonary valvotomy at age 5
26	11 years	F	anomalous subclavian artery, hypoplastic descending aorta, PS, bilateral PAS (hypoplastic & tortuous pulmonary arteries)	30/9	17/9	.....		.....
27	22 years	M	Bilateral PAS	70/18	43/20	30/15		.....
28	7 years	M	PAS	32/6	21/6	.....		.....
29	3 years	M	VSD, PAS	80/32	58/25	.....	.....	VSD repaired at age 3
30	2 years	M	Bilateral PAS (Hypoplastic)	100/13	15/8	15/5		Died at operation at age 2
31	1 year	F	VSD, PS, bilateral PAS	30/18	23/15	20/13		.....
32	11 years	F	VSD, PS, bilateral PAS	30/12	18/11	22/10		.....

branches. The incidence of this condition in the present series was 2.7 per cent (22 to 821 patients). The remarkable observation in this series is the overlap between these groups, making it possible to say that 50 per cent of the patients with congenital rubella syndrome were found to have pulmonary artery stenosis as some part of their cardiac malformation and that among patients found to have pulmonary artery stenosis almost half had congenital rubella syndrome.

Rowe's<sup>6</sup> remarks on this association sprang from an experience with a rubella epidemic in New Zealand. In his report published in 1963<sup>8</sup> he discussed 35 patients with congenital rubella syndrome, 11 of whom had pulmonary artery stenosis. As the obverse, he also mentioned 18 patients with pulmonary artery stenosis, in 11 of whom the lesion was associated with congenital rubella. Since specific questioning about maternal rubella infection was part of history taking in the San Diego County Heart Center during the eight-year period reviewed, the fact that the majority of the patients in the series are young is due to a similar epidemic of rubella that occurred in many parts of this country in recent years.

Why rubella virus has such a predilection for the structure of the embryologic sixth dorsal arch is not known, but it is certainly true that patent ductus arteriosus and pulmonary artery stenosis far exceed valvular and septal defects in this infection.

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